Parents require ongoing supportive casework to cope with long-term responsibility and care, to make decisions repeatedly about hospitalization and procedures. They need encouragement to maintain social activities away from the child and to ask for and find resources for relief from 24-hour care. Fifteen-year-old Ted had massive orthopedic involvement. His mother was a single parent who until his first clinic visit had hardly left the house for several years. Parents are greatly in need of genetic counseling to resolve feelings of guilt which are often present and to fully understand the risk of recurrence of spina bifida in future children.

The social worker may have to emphasize over and over the value in a child's being as independent as possible. Two years went by before ten-year-old Mary's mother reached a point where she could allow Mary to change her own diapers. The older child who presents a physically normal appearance but lacks bowel and bladder control needs not only adequate devices and instruction in self-care but help in accepting and living with this problem. The focus of casework is on developing an optimal self-image. It was only when Jane, 13, began to feel some sense of self-worth that she was ready to keep herself clean enough to eliminate the odor of urine which had made her open to ridicule and social isolation.

Psychosexual counseling is crucial for teenagers and parents. The older the child, the more specific the information he or she must be given. To provide this type of counseling the social worker has to be well informed and comfortable in dealing with the subject. Tom, 18, who wore a catheter, was starting college without ever having dated and had decided he would never marry. In one clinic visit with the social worker he began to see the possibilities open to him for achieving an adequate sexual relationship and to approach the subject of his psychosexual functioning from a different viewpoint.

The social worker is an important link in the chain of communication between child and family and clinic team. She informs the team of psychosocial problems which interfere with treatment or make it unrealistic to expect that recommendations will be carried out. Paul was confined to a wheelchair and was obese. The team repeatedly urged his mother to put him on a diet (which she did not do) until the social worker pointed out that the whole family was obese and not apt to change their eating habits. The social worker is sometimes the only member of the team who knows all the questions the child or family has, so she makes sure that questions are asked and answers are understood. She is usually the liaison who keeps communication open between clinic team and community agencies. The importance of her role in maintaining the flow of communication in all directions cannot be overemphasized.

Refer to: Pevehouse BC: Myelomeningocele—Part V: Neurosurgical aspects, *In* Myelomeningocele—A symposium. West J Med 121:298-300, Oct 1974

MYELOMENINGOCELE—PART V

Neurosurgical Aspects

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THE PROBLEMS of myelomeningocele and meningocele are those of spinal dysraphism. These are not isolated defects, but rather conditions that involve the entire motor system. The ventricles and the circulation of the spinal fluid, as well as the spinal cord and its coverings, are involved. The meningocele that one observes is actually an outline of the meninges protruding through a bony defect; it has spinal fluid in it and occasionally it will also have some fat but it has no nerve tissue, this absence being basic to the definition of a meningocele—that is, the cord and the nerve roots are intact and are not involved in the defect.

A more serious lesion, that of meningomyelocele, or what we now prefer to call myelomeningocele, putting the cord part first, always involves the spinal cord as a part of the defect. There may be displacement of the cord, duplication of the cord, or separation of it, often in the lumbosacral region, and there will be tethering of roots in the dome of the defect with a consequent neurological deficit. We have found the ratio of myelomeningocele to simple meningocele to be about eight to one. There are also more rare and complicated forms of rachischisis in which the cord is displaced into

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the defect or split, and in which there is dilatation of the central canal. These lesions all have an individuality of their own. No two defects look exactly the same; they come in all sizes, shapes, and locations.

The Arnold-Chiari malformation is very similar. It is a part of the whole problem of spina bifida in that there is deformity of the ventricles, the midbrain, and the posterior structures of the cerebellum, resulting in displacement of the cerebellar tonsils downward into the cervical canal and compression of the cord, which in turn causes obstruction of spinal fluid flow through the area. Displacement of the medulla causes disturbances of the lower cranial nerves, and any child with this deformity may have trouble breathing, or stridor when crying. Such a child is in danger of having respiratory arrest.

Other lesions that occur may be occult but are often serious. Lipomas involving the cord or roots may be heralded only by a small tuft of hair at the base of the lumbar spine or a dimple which in some instances will be a source of repeated meningitis, and not until the second or third episode in the same child—following which there may be serious deficits as a result of the meningitis—does the reason become apparent.

It should again be emphasized how many lesions are in the lumbosacral area. In our series of 295 lesions there were only seven above the level of the first lumbar vertebra. They may, however, occur anywhere along the neural tube. The defect has its origin about the thirtieth to forty-fifth day of gestation with failure of closure of the neural tube. It may occur at the very rostral end and involve the cribriform plate and the base of the brain, or it may be anywhere as far caudally as the sacrum.

Even in what may appear to be a simple meningocele, there may be some neurological deficit indicating that the lesion is in reality a myelomemingocele. As the child grows a little older, some lesions may develop into a cicatrix following a granular repair within a lesion. One may see abnormal hair in some. Another feature of the myelomeningocele sac is the abnormal skin around the periphery with a very attenuated arachnoid in the center, directly beneath which lies spinal fluid. This is the area from which leakage takes place, and leakage is the great danger since it may lead to retrograde infection and meningitis.

Some of the lesions of the cranium can also cause ventricular displacement in which there may be a connection between the third ventricle and the two lateral ventricles, often accompanied by distortion of the cerebellum and occasionally with involvement of some of the venous sinuses that are part of the tentorium. As mentioned above, however, the typical defect is lumbosacral and is accompanied by a neurological defect. There is usually both bowel and bladder disturbance. In many of these babies rectal prolapse develops very rapidly, because of sphincter incompetence, and the prolapsed segment may feed out 15, 20, or 25 cm of mucosa. This prolapsed segment has to be protected during the first few weeks in many of the children, and to do so the buttocks are often taped together.

In our efforts to preserve whatever neurological function is present, we are concerned with the neural tissue in the center of the sac. The neural elements are usually extruded and must be covered in order to prevent infection, by constructing a layer of meninges which will prevent spinal fluid leak and allow some sort of satisfactory skin closure. These children often have problems with leakage of spinal fluid because 80 to 90 percent of them have hydrocephalus resulting from the dysraphism itself. Some of them have communicating hydrocephalus, some have aqueductal blocks, or in the case of the Arnold-Chiari malformation there may be a block of the foramen magnum. In the repair of these lesions it is important to identify the neural elements, although the cord may end in the defect, with no continuation; such a child is totally paraplegic. In this event it is important to tailor the neural elements properly to prevent tethering of the cord as the child grows, and also to effect a proper dural closure to keep the spinal fluid system intact. Sutures in the skin are avoided whenever possible unless there is tension on the grafts. Mere removal of these lesions greatly simplifies nursing care. There are children who have gone as long as three years with large lesions, but from a technical standpoint 98 percent of all lesions of this sort can be closed. It has not been clearly demonstrated that this makes any great difference neurologically, but it is our belief that by preventing complicating meningitis, and also by trying to preserve all the remaining neural elements that are present, there may be some sensory reinnervation to a level or two lower at some later time. There are a number of patients who have not been operated upon and have survived; in this type of series one finds that such children had small le-

sions which spontaneously granulated in. Thus they did not require operation and still survived. However, nearly all children not operated upon will die. If the primary physician, or other personnel involved, advises against repair of the lesions, then there must be some method of euthanasia so that such children do not constitute a burden to either the parents or the state.

After the spinal lesion has been closed and the problems of meningitis are gone, the neurosurgical problem that then remains is control of the hydrocephalus which usually develops. The onset of hydrocephalus in the newborn is apparent either at birth or within the first month or so. This is important for the primary physician to realize. The head should be measured, the fontanelle should be inspected and palpated to determine tenseness, and the findings should be recorded so that a week or two weeks later it can be determined whether there has been a change. Many times a baby is seen at one or two months of age and there is a question as to whether hydrocephalus exists or not, but there is no previous head circumference measurement available for comparison, even though the ventricles are in fact beginning to dilate. As far as the remainder of the neural tube is concerned, there may be associated defects anywhere which can cause hydrocephalus; they can be at the aqueduct, as is often the case in the Arnold-Chiari malformation, or in the posterior fossa. Occasionally, failure of development of the proper system of arachnoidal villi to return spinal fluid to the blood circulation is postulated. A block anywhere in the circulation of cerebrospinal fluid will produce hydrocephalus, and anything which obstructs the one-way flow of spinal fluid as it enters the venous system does the same thing although more slowly.

There are many terms, such as communicating hydrocephalus (so called because a dye placed in the ventricular system can be recovered in the spinal fluid), but that does not necessarily mean that there is no obstruction around the base of the system, such as in a postmeningitis situation. Any block, depending on its location, may cause enlargement of the head from increased intracranial pressure. It is obvious that a block of the aqueduct will cause a very rapid ballooning of the ventricles. A block around the base of the system may be partial, so that head growth may be only very slightly increased over normal, and it may take a long time to make the diagnosis that the ventricles are abnormally enlarged.

The present method of treatment is to place a shunt that goes from the ventricular system either to the peritoneum or to the blood stream through the jugular vein, reaching the auricle via the superior vena cava. Several types of shunts are available, some with a palpable reservoir, placed in a bur hole, that provides a little pumping system. When a shunt is made in a child with spina bifida, the meningocele itself in many instances will collapse and make it easier to treat, but there is a converse problem in that if infection is present, it sometimes will get into the system and thereby cause the very complication that it is intended to avoid, namely, infection of the shunt itself, with subsequent obstruction to spinal fluid flow. The decision regarding therapy is a complex problem; each case must be dealt with by first making an accurate evaluation of flow and then trying to see which approach appears best. In many instances a shunt is not the best answer for a child with minimal hydrocephalus. There are many reasons for this, relating of course to the possibility of postoperative infection and also to the effects on the ventricular wall in a marginal hydrocephalus that may already be arrested. Problems often occur in maintaining proper function of shunts, but if we can control the hydrocephalus after the lesion has been repaired, then the real threat to life is urinary tract infection.

Refer to: Cohen P: Myelomeningocele—Part VI: General considerations, *In* Myelomeningocele—A symposium. West J Med 121:300-304, Oct 1974

MYELOMENINGOCELE—PART VI

General Considerations

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SPINA BIFIDA is a disease entity which includes a group of developmental defects of the spinal column in which there is failure of fusion of vertebral arches, with or without protrusion and dysplasia

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